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Case Report

A case report of exhibitionist behaviour revealing unsuspected Huntington's disease: A pitfall in forensic medicine

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Abstract

We present the case of a patient who was recently convicted for exhibitionism whose clinical assessment suggested undiagnosed Huntington's disease. We summarise the clinical characteristics, the genetics involved and the diagnostic difficulties which can be encountered. Finally, we discuss the implications of this diagnosis in forensic medicine.

Keywords: Huntington's disease; Chorea; Exhibitionism; Forensic medicine

1. Introduction

It is often crucial to be able to provide proof that reprehensible acts are a consequence of a qualified disease, particularly when the legal responsibility of the perpetrator is in question. In most instances, the relationship can be easily established, for example, when mental disorders or severe neurological impairment are present... In a few cases however, it is much more difficult and subtle signs related to rare diseases need to be sought. We present the case of a patient who was recently convicted for exhibitionism whose clinical assessment suggested theretofore undiagnosed Huntington's disease. We summarise the clinical characteristics, the genetics involved and the diagnostic difficulties which can be encountered. Finally, we discuss the implications of this diagnosis in forensic medicine.

2. Observation

A 69-years-old retired man was referred to our neurological consultation by his psychiatrist for gait disturbance and a lack of introspection concerning his personal bearing, suggesting cognitive impairment. The patient had been convicted three times since 2001 for sexual exhibitionism and had been released on parole one month prior to being seen in our clinic. He was legally required to undergo psychotherapy. Because of a recent new offence, a new conviction was likely. On neurological examination, the patient demonstrated intermittent, jerky movements in both hands and legs during walking, accompanied by oro-facial dyskinesia and motor impersistence. The cognitive examination revealed that the patient was fully oriented in time and space but had trouble following instructions and had

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difficulties in shifting abilities and working memory, suggesting frontal dysfunction.

The clinical picture suggested Huntington's disease (HD) and a genetic analysis were performed. This showed abnormal repetition of CAG triplets on both alleles of the Huntington gene: 40 repetitions on the first allele and 28 repetitions on the second one.

3. Discussion

HD is a neurodegenerative disease with an insidious onset associating motor signs, cognitive decline and psychiatric disturbances.[1] This symptomatic triad can be incomplete, especially at the onset of the disease.[2] Motor signs consist in rapid, irregular, uncontrolled and involuntary jerky movements of the limbs that interfere with normal motor activity. Cognitive decline is characterised by precocious frontal dysfunction with impairment of the executive functions such as planning, organising, anticipating the consequences of actions and eventually leading to dementia when cognition is massively impaired. The psychiatric disturbances consist in depressed mood (with very frequent suicidal ideations), dysinhibition, irritability, apathy, manic or psychotic episodes[3]... Behavioural disorders are directly related to functional decline but their occurrence appears to be independent of cognitive or motor dysfunction.[4]

HD has an autosomal dominant transmission. The HD gene is located on the short arm of chromosome 4 and is associated with an excessive number of trinucleotide repeats. The disease exhibits full penetration when CAG repeats reach 41 and incomplete penetration when it repeats between 36 and 40. Trinucleotide repeats that exceed 28 show instability on replication, usually leading to expansion from generation to generation. Thus, new-onset cases of Huntington's disease with a negative family history can appear, as in our observation. Our case report is unusual because the abnormal repetition of CAG triplets on both alleles suggests mutation or premutation in both parents' genotypes.

Disturbances in sexual behaviour are infrequent in HD, but classically mentioned.[5] Hyposexuality is the most frequent disturbance encountered[5]; hypersexuality is observed in less than 5% of the patients[1] and dysinhibition can intensify sexual impulses. Exceptionally, some of patients present sexual deviance.[6]

HD should be systematically ruled-out as a possible cause of behavioural disturbances and/or abnormal sexual behaviour in patients, whatever the age of onset or family history. Moreover, some of the symptoms of the clinical triad can be prominent[2] and an incomplete pattern requires genetic studies in order to confirm the diagnosis. The consequences for patients, their relatives and the implications in forensic medicine are crucial.

4. Financial Disclosure and Sponsor's Role

None.

Conflict of Interest

None.

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